

Radiology Corner

Chondroblastoma with Secondary Aneurysmal Bone Cyst

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Note: This is the full text version of the radiology corner question published in the February 2008 issue, with the abbreviated answer in the March 2008 issue.

We present the case of a chondroblastoma with secondary aneurysmal bone cyst arising in a posterior rib. The tumor was initially detected on a chest radiograph obtained for evaluation of chest wall pain. This case illustrates the importance of a systematic search pattern in evaluation of chest radiographs. The following report reviews the clinical, radiologic, and pathologic characteristics of primary bone tumors and presents a differential diagnosis for posterior mediastinal mass, then paraspinal osseous mass.

Introduction

A 24 year old male presents to his provider with right-sided, lower posterior chest wall rib pain for approximately 4-6 months, which has been increasing in severity. He notes that the pain increases with deep inspiration. He denies a history of trauma in the area. On physical exam, right paraspinal tenderness upon palpation is present, approximated at the 9th posterior rib.

Summary of Imaging Findings

The PA chest x-ray (fig. 1) demonstrates a large opacity visible in right lung field at level of right hilum. The density has fairly well delineated borders superiorly and inferiorly, however, is less well defined laterally. The opacity does not silhouette the right heart border and the hilar vessels are not obliterated (likely placing this mass outside the middle mediastinum, and most likely posterior). On closer inspection, the lateral margin of the 9th vertebral body is indistinct. Also, the medial aspect of the posterior right 9th rib is not well defined, supporting a posterior mediastinal or chest wall (ribs, soft tissue) location.

On the lateral chest (fig. 2) the mass is confirmed to be posterior. Note the positive "spine sign" in that the thoracic spine does not get progressively darker the more inferior one

looks. Also note the obtuse angles of the margins of the mass, supporting chest wall or pleural location.

These findings are better seen on the AP radiograph of the thoracic spine (fig. 3). Coronal reconstructed CT image (fig. 4) displays the lesion arising from the right posterior 9th rib. Axial CT images (fig. 5) demonstrate invasion of the right T9 pedicle and destruction/expansion of the right posterior 9th rib.

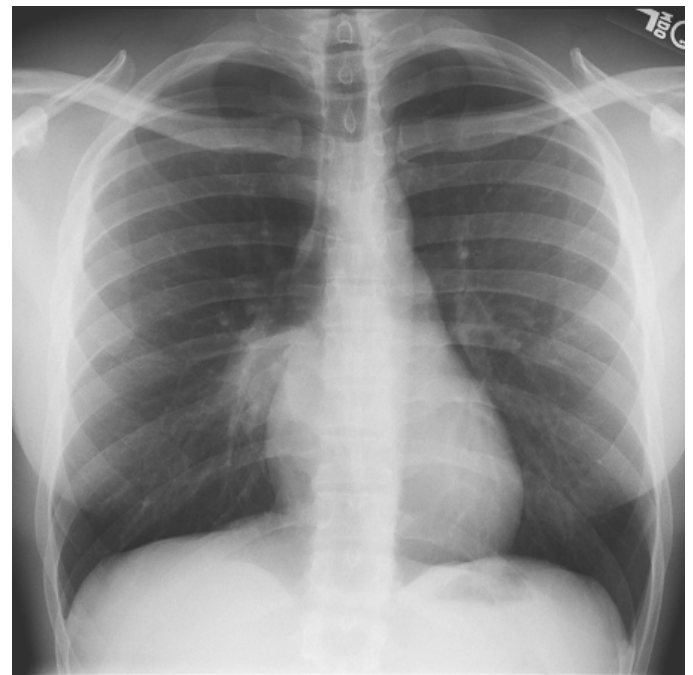


Fig. 1. Large opacity noted in right lung field, at level of right hilum. The density has fairly well delineated borders superiorly and inferiorly, but becomes less well defined laterally. Lesion does not silhouette the right heart border and does not obliterate hilar vasculature. The 9th rib margins adjacent to thoracic spine are poorly defined, as well as the lateral margin of the T9 vertebral body.

Axial T1W MR images with and without contrast (figs. 6 and 7) demonstrate a large, lobulated mass located in right paraspinal region, precisely at the costovertebral junction of T9 which appears to have epicenter at the ninth rib. Mass extends into the neural foramen and abuts the thecal sac; it measures 6.9x4.7x4.6 cm. Post-gadolinium image shows mild enhancement with multiple areas of low signal within the mass. Axial T2W MR (fig. 8) shows multiple fluid-fluid levels. Coronal T2W MR (fig. 9) demonstrates right paraspinal mass spanning levels T8-T10, with invasion of the right pedicle of T9.

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Patient Discussion

Considering the patient's demographics, the radiographic features, and the confirmed histologic presence of cartilage and osteoblasts; there are several neoplasms included in the differential. Likely neoplasms include several benign primary bone tumors, malignant primary bone tumors, as well as neoplasms of alternate tissue origin, such as aneurysmal bone cysts, neurogenic tumors, and expansile metastatic lesions such as thyroid or renal cell carcinoma.¹ Probable benign tumors include: fibrous dysplasia, chondroblastoma, chondromyxoid fibroma, osteochondroma, giant cell tumors, or enchondroma. Chondrosarcoma, osteoblastoma—aggressive variant and osteosarcoma represent the likely malignant neoplasms.^{2,4} The final diagnosis in our case is a chondroblastoma with secondary aneurysmal bone cyst, confirmed by core biopsy and pathology.

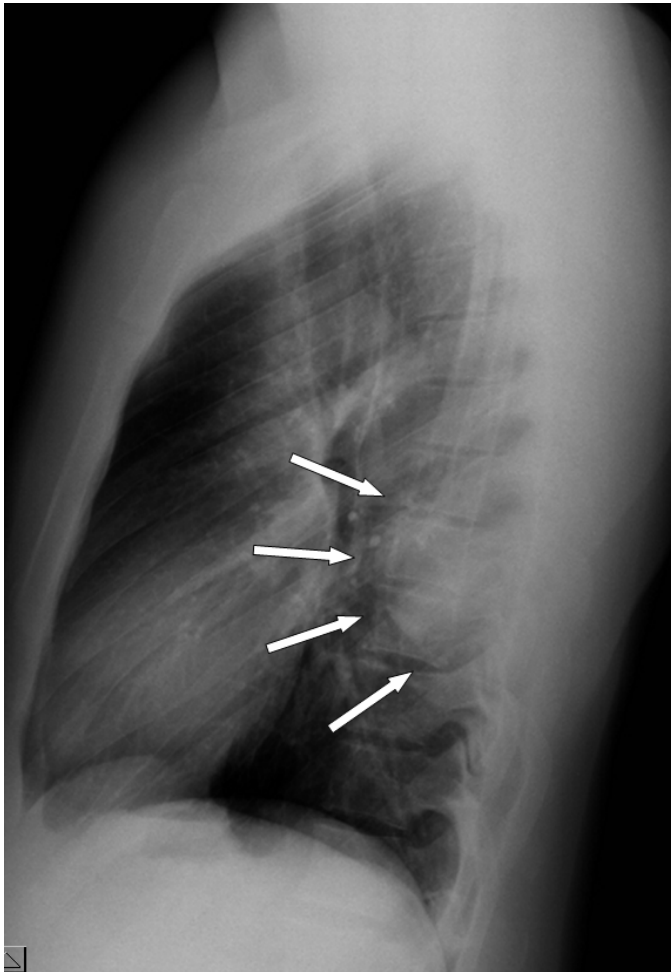


Fig. 2: Lateral chest radiograph demonstrates density overlying thoracic spine, approximated at levels T6-T8. Inferior margin is likely obtuse, though margins superior are poorly visualized (arrows).

Discussion

The following describes some familiar features of tumors of the chondro-osseous origin, with mean age of presentation,

most common anatomic location, likely radiographic findings, and histologic correlation given for each tumor.

Chondroblastomas are neoplasms of the chondroid matrix that occur most frequently in the second decade of life. The most common location of these tumors is in the long bones, with the femur having the highest incidence. Radiologically the mass appears lobulated with scalloped cortical margins. Calcifications and cysts are often described.^{2,3,5}

Chondromyxoid fibromas present in the second and third decades, with predominant location in the lower extremities. The metaphyses of the tibiae and fibulae are likely to be involved. Plain films will demonstrate an eccentric lytic lesion and T1W MR shows decreased signal. Histology displays a chondroid matrix with cytologic features of anaplastic processes.^{2,7}

Osteochondromas are benign neoplasms composed of osteoid and cartilage that occur from adolescence to the early 20's. Often described as occurring "around the knee," they tend to be located in long bones. Exophytic lesions are sometimes seen on radiographs, but usually they are subperiosteal.^{2,7}

Giant cell tumors of the bone present in patients 25-40 years of age. Another tumor of the long bones, these lesions are located in upper and lower extremities. Radiographically they tend to be solitary, locally aggressive, and highly expansile.²

Enchondromas also occur in the long bones, often in the third decade. Lesions tend to be oval, lytic with scalloped cortices on imaging. Histology reveals cartilage with a layer of smooth, thin bone surrounding.²

Chondrosarcomas tend to occur in an older population than most primary bone tumors, with peak incidence in the 5th and 6th decades. Femur, humerus, ribs, and pelvis are the most commonly cited locations. Imaging shows lucenies, calcifications, scalloped edges, and extension into soft tissues. Being a malignant neoplasm, pathology exhibits cartilage with anaplasia.^{2,7}

Osteoblastoma, aggressive variant, is often considered a malignant lesion (though technically categorized as benign) because of its impressive recurrence potential. Pathology shows osteoid, osteoblasts, spindle cells, with rare cartilage. Like other primary bone tumors, they often occur in the second decade, though unique is its location which is in the vertebra and posterior spinal elements. Radiographic characteristics vary greatly with this tumor from lucent to sclerotic, confined to expansile, and often there is adjacent bone thinning.^{2,7}

Osteosarcomas are composed of anaplastic osteoblasts, chondroblasts, and fibroblasts. Location tends to center around the long bones of the knee and the neoplasms occur from adolescence up to the third decade. Radiographs of these tumors vary from lytic to sclerotic, to mixed variants.^{2,7}

Treatment and Follow-up:

After US-guided core biopsies were obtained, metastatic work-up was completed, including whole-body bone scan, FDG PET scan, chest CT, and brain MRI. There was no evidence for metastatic disease.

The tumor was resected en bloc. Final pathologic evaluation of the resected specimen confirmed the diagnosis of chondroblastoma. Despite their benign taxonomy, a majority of these benign lesions tend to recur, therefore making wide excision standard regardless of malignant potential.⁶

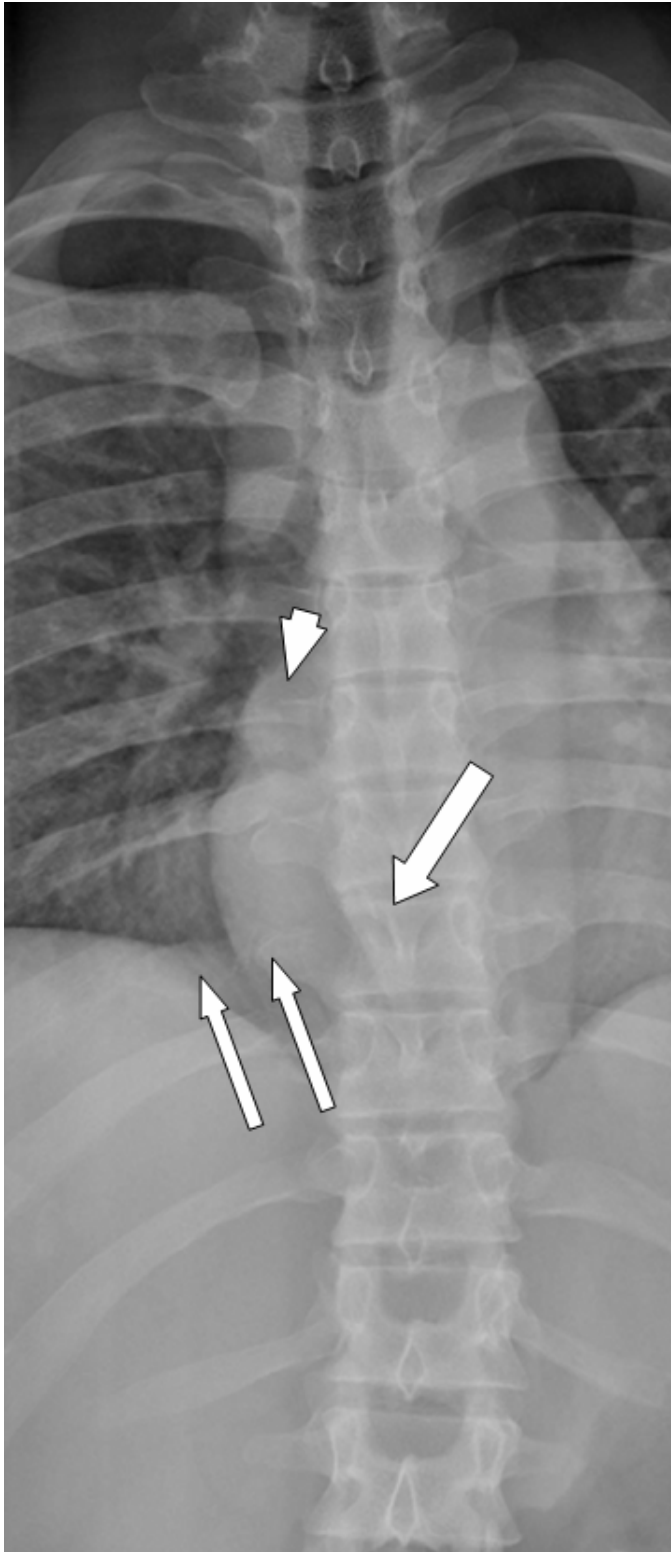


Fig. 3: AP Thoracic spine demonstrates multiple bony erosions associated with right paraspinal mass: right transverse process of T7 (arrowhead), T9 vertebral body and right pedicle (wide arrow), and the medial 5 cm of right 9th rib (arrows). This plain radiographic modality helped narrow the differential diagnosis significantly in that we are now dealing with an osseous (or less likely an aggressive pleural) process.



Fig. 4: Coronal CT with contrast (IV): Coronal reconstructed image displays lesion arising from the 9th rib, posteriorly.

Paraspinal masses invoke an enormous list of differential diagnoses: lymphoma, metastatic disease, nerve sheath tumors, primary bone tumors, myeloma, meningocele, cysts, abscesses, and beyond. To narrow this vast list of potential diagnoses, diagnostic clues, such as location, radiographic features, and patient age should be carefully scrutinized. Based on these parameters, with radiologic features of utmost importance, this particular patient's findings suggested a neoplastic process, which directed him to biopsy. Indeed biopsy results initially indicated that a primary bone neoplasm was credible, based on presence of osteoblasts and cartilage.

Primary bone tumors are relatively rare, with an incidence of approximately 10,000 cases a year in the US.^{4,7} Using the patient's demographics, radiographic features, and the confirmed presence of cartilage and osteoblasts, the differential above mentioned was considered. All of those suspected neoplasms, with the exception of chondrosarcoma, fit this patient's age group, as all frequently present in the second and third decades (chondrosarcoma has a peak incidence in the 5th and 6th decades of life). Using location of the mass to assign a probable diagnosis, an osteoblastoma

becomes likely as it is often found in the spinal elements. The intrinsic characteristics of the mass on MR imaging indicate an aneurysmal bone cyst component to the tumor (due to the presence of fluid-fluid levels). Because only a small fraction of the tumor contains these structures, they are likely to be secondary rather than a primary aneurysmal bone cyst.

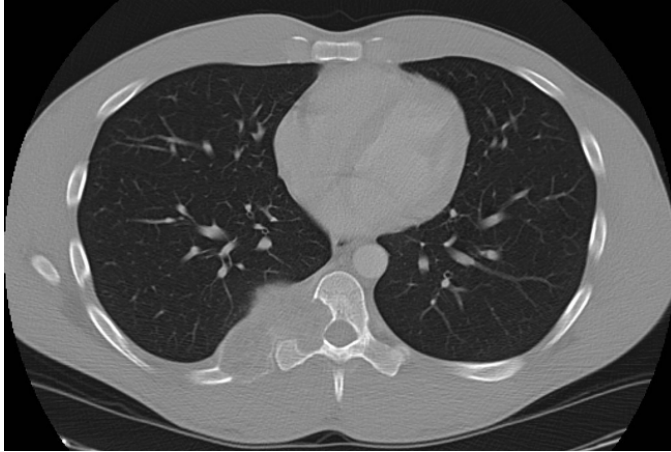


Fig 5. Axial : CT w/contrast (IV). Large, lobulated mass located in right paraspinal region. Representative slice is at T9 and shows bony destruction of the T9 vertebral body and pedicle, as well as erosion of the ninth rib. Subsequent slices demonstrate involved of T8, T9, and T10; mass extension into neural foramina; no hilar or mediastinal pathologic lymphadenopathy is present.

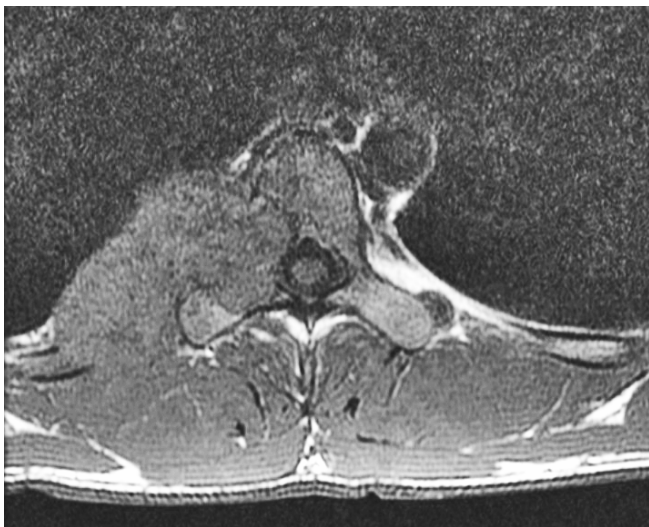


Fig. 6. Axial : MR - T1W – noncontrast: Large, lobulated mass located in right paraspinal region, precisely at the costovertebral junction of T9 that appears to originate from the ninth rib. Mass extends into the neural foramen and abuts the thecal sac. Mass measures 6.9x4.7x4.6 cm.

When considering primary bone tumors with secondary aneurysmal bone cyst components, the differential is limited. Only five types of primary bone tumors typically display this pattern: giant cell tumor of bone, osteoblastoma, chondroblastoma, fibrous dysplasia, and telangiectatic osteosarcoma.^{4,7} Final pathology reading of the resected specimen revealed a diagnosis of chondroblastoma with areas of secondary aneurysmal bone cyst.

This is a very rare tumor, and even rarer is the location of the tumor in this patient. Chondroblastomas account for less than 1% of all bone tumors; this corresponds to approximately 100 cases a year, considering the scarcity of primary bone tumors in general. Additionally, nearly 75% of the cases occur in the long bones of the lower extremity - distal femur and proximal tibia/fibula. Another 20% of chondroblastomas occur in the humerus, while a small fraction have been reported in the small bones of the hand and feet.^{2,3,5,7} Case reports have described these tumors occurring in the skull, mandible, maxilla, vertebra, ribs, scapula, patella, and sternum.¹ Osteoblastoma was excluded pathologically by the presence of cartilage within the tumor, and by immunohistochemical staining positive for S-100.^{2,8} Areas of reactive osteoid were present, but no osteoid producing tumor cells were seen.



Fig. 7. Axial : MR - T1W w/Gadolinium: Large, lobulated mass located in right paraspinal region, precisely at the costovertebral junction of T9 that appears to originate from the ninth rib. Mass extends into the neural foramen and abuts the thecal sac; measures 6.9x4.7x4.6 cm. Post gadolinium administration shows mild enhancement with multiple areas of low signal within the mass.

This patient fits the expected demographics of the majority of primary bone tumors: second decade in age, male, black, with history of chronic pain.^{4,7} What is so profound regarding this case remains the specific type of primary bone tumor, given its rarity by statistical incidence, and its unusual location originating from the ninth rib.^{2,3,5,7}



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Fig 8. Axial : MR - T2W. Large, lobulated mass located in right paraspinal region, at the costovertebral junction of T9. The mass invades the right pedicle of T9 and extends into the neural foramen, where it abuts the thecal sac. Multiple fluid-fluid levels are present (arrows).



Fig. 9. Coronal : MR - T2W. T2W coronal image demonstrates right paraspinal heterogeneous mass spanning levels T8-T10, with invasion of the right pedicle of T9.

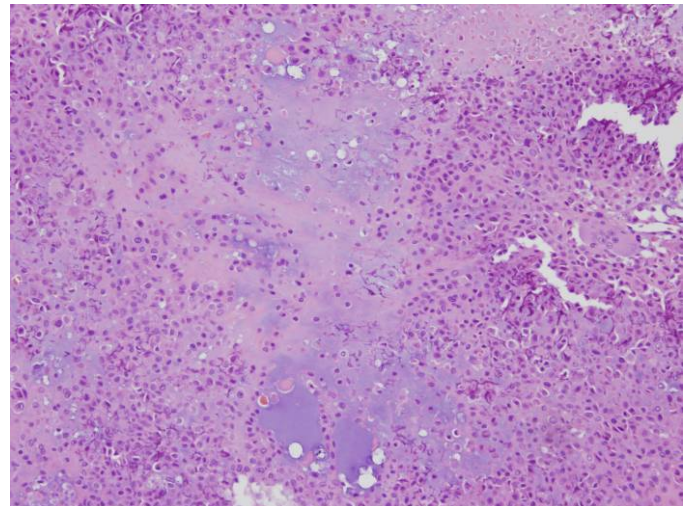


Fig 10: Photomicrograph (original magnification, x400; hematoxylin-eosin stain) shows sheets of relatively uniform medium sized cells surrounding areas of bluish hyaline cartilage and more eosinophilic cartilage matrix.

Category 1 CME or CNE can be obtained on this case in the MedPix™ digital teaching file on the following link:

<http://rad.usuhs.mil/amsus.html>

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